Wunderlich's Syndrome in a Tuberous Sclerosis Patient

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Abstract Spontaneous non-traumatic renal hemorrhage known as Wunderlich's syndrome is known to occur in renal angiomyolipoma and may be the first manifestation of the disease. Angiomyolipomas in tuberous sclerosis are usually bilateral and multicentric. A 25yr old female, a case of Tuberous sclerosis with bilateral renal angiomyolipomas presented with right loin pain of increasing severity. On evaluation she had acute abdomen with increasing abdominal distension. Investigations revealed large right perinephric hematoma with bilateral renal angiomyolipomas. Exploratory laparotomy and right nephrectomy was done. Spontaneous hemorrhage in renal angiomyolipoma in a case of tuberous sclerosis and management are discussed.

Keywords Tuberous sclerosis syndrome (TS) · Renal angiomyolipomas · Wunderlich's syndrome

Introduction

Wunderlich's syndrome, a urological emergency, refers to spontaneous nontraumatic renal bleeding confined to the subcapsular and/or perinephric space. Various etiologies include benign and malignant renal neoplasms, vascular disease (vasculitis, renal artery arteriosclerosis, and renal artery aneurysm rupture), nephritis, infections, undiagnosed hematological disorders, and anatomical lesions. Of all

these etiologies, the commonest cause in most series is renal angiomyolipoma (AML).

Angiomyolipoma is a benign clonal neoplasm consisting of varying amounts of mature adipose tissue, smooth muscle, and thick-walled vessels within renal parenchyma. It is seen in more than 50% of patients with tuberous sclerosis syndrome (TS) [1]. AML in TS is more likely bilateral and multicentric, and has a tendency toward accelerated growth. We report a patient of TS who presented with spontaneous retroperitoneal hemorrhage due to renal AML. Massive retroperitoneal hemorrhage from AMLs, also known as Wunderlich's syndrome, represents the most significant and feared complication.

Case Report

A 25-year-old female is presented with right loin pain of oneweek duration. On examination, it was found that she had adenoma sebaceum over face (Fig. 1) and mild tenderness in right lumbar region. Following investigations, ultrasound of the abdomen and contrast enhanced CT scan of the abdomen revealed bilateral enlarged kidneys with multicentric nodules with fat densities (<20 HU) within both the renal parenchyma, confirming the diagnosis of bilateral renal AMLs. CT brain showed subependymal calcification. This pattern of cerebral calcification together with adenoma sebaceum was diagnostic of TS, she was diagnosed as a case of TS with bilateral renal AMLs and was conservatively managed.

The same patient was presented six months later with severe right loin pain. On examination, she was pale and abdomen was distended with tenderness and guarding of right lumbar and iliac regions. CECT scan of abdomen showed massive right perinephric hematoma and bilateral renal AMLs (Fig. 2). In view of expanding retroperitoneal hematoma, emergency laparotomy and right nephrectomy was planned.

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Fig. 1 Tuberous sclerosis patient with adenoma sebaceum over the face

Bilateral subcostal (chevron) incision was made and right kidney was exposed transperitoneally by reflecting the right colon and hepatic flexure by Cantrallization. Duodenum was kocherized to expose hilum of right kidney. Owing to huge hematoma crossing midline, technical difficulty of approaching the hilum was faced. Ongoing bleeding from the upper pole was temporarily contained by pressure packs while renal pedicle was dissected and clamped with Satinsky vascular clamp. Hematomas were partially evacuated to facilitate exposure of renal hilum. After ligating the renal artery and renal vein, kidney along with the hematoma was removed. Postoperatively patient has normal serum creatinine and is on regular follow-up.

Discussion

Angiomyolipomas are known to occur in TS, an autosomal dominant disorder characterized by mental retardation, epilepsy, and adenoma sebaceum, a distinctive skin lesion [1]. The growth of these tumours is thought to be hormone dependent as suggested by female predominance and rarity before puberty. AML in TS is usually bilateral and multicentric.

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Clinical presentation of Wunderlich's syndrome depends on the degree and the duration of the bleeding. The classic presentation includes flank or abdominal pain, a palpable

tender mass, and gross hematuria (Lenk's triad) [2]. Other symptoms and/or signs are nausea, vomiting, fever, anemia, renal failure, and hypotension. The syndrome can be fatal if not identified and promptly treated. Pregnancy increases the risk of bleeding in AML. Sporadic reports of massive retroperitoneal hemorrhage and shock during pregnancy have been described [3]. Longstanding perinephric hematoma can compress the renal parenchyma and lead to hypertension and deterioration of the renal function, a condition called page kidney. CT scan is the investigative modality of choice in diagnosing AML and its related complications. On ultrasound, AML is typically intensely echogenic and can cause acoustic shadowing. Presence of fat in a noncalcified renal mass lesion on CT confirms the diagnosis of AML in most cases [4].

The management of Wunderlich's syndrome depends on the general condition of the patient. In patients with solitary tumour or tumours confined to one part of the kidney, angioembolization can arrest the bleeding and these patients can later be considered for a nephron sparing surgery in the form of partial nephrectomy. Selective embolization and nephron sparing surgery should be considered especially in patients with bilateral disease and pre-existing renal insufficiency. However, coalescent renal AML replacing entire renal parenchyma is a much more vexing clinical problem. When bleeding occurs in this circumstance, it can be impossible to identify which lesion is the source. Nephron sparing surgery is not feasible when entire renal parenchyma is replaced by multiple tumours. The role of embolization in the management of existing or impending hemorrhage is expanding [5]. Both embolization and nephron-sparing surgery have been reported with excellent

Fig. 2 CECT scan of abdomen showing bilateral renal angiomyolipomas with large right perinephric hematoma

success and minimal complications. Embolization can temporize and in many cases, arrest bleeding. A nephron-sparing approach in the form of partial nephrectomy should be considered when possible particularly in patients where preservation of renal function is at issue [6]. Patients with life-threatening hemorrhage, failed embolization require urgent exploration and in majority of the cases require total nephrectomy [7]. We considered nephrectomy as our patient was hemodynamically unstable.

The management of perirenal hemorrhage in pregnancy depends on the degree and duration of bleeding, clinical condition of the patient, and duration of pregnancy [3]. Patients with nonexpanding hematoma and stable clinical status in first trimester of pregnancy can be conservatively managed till second trimester. Angioembolization is the preferred mode of treatment to control the bleeding in pregnancy. However, it carries risk of radiation exposure to the fetus. If angioembolization is required to stop the bleeding in first trimester, medical termination of pregnancy should be strongly considered. In second trimester, angioembolization can be performed with brief exposure to fluoroscopy during the procedure. Coalescent renal angiomyolipomata replacing entire renal parenchyma is a much more vexing clinical problem. When bleeding occurs in this circumstance, it can be impossible to identify which lesion is the source [8].

As the renal AML was bilateral in our patient, she is theoretically prone for Wunderlich syndrome on the contralateral side also. The patient and her relatives are well

educated about the nature of pathology. She is informed to report immediately in case of sudden abdominal pain and/or hematuria. We would consider angio-embolization and nephron sparing surgery if she develops contralateral bleed. She was also educated as to the possible need of renal transplantation should she require contralateral nephrectomy owing to failed embolization or torrential bleed.

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